MEDICO-LEGAL

MEDICO-LEGAL ASPECTS OF INTERSEXUALITY: CRITERIA OF SEX*

KEITH L. MOORE, Ph.D.† and
CLIFFORD H. C. EDWARDS, LL.B.,‡
Winnipeg, Man.

PART I OF TWO PARTS

RELATIVELY RECENT developments concerning the criteria of sex have raised questions that have medico-legal implications. Society and the law recognize only two sexes. Usually the differences are obvious, partly because cosmetic and garment manufacturers make it easy for females to accentuate or falsify their characteristics.

At birth the physician assigns sex from the appearance of the external genitalia. If these organs are equivocal, other criteria must be used. In the past a final decision was usually not made until the type of gonads had been determined. Often this was not done until contradictory sex characteristics appeared at puberty; frequently in such cases sex was changed, resulting many times in unhappy confused patients.

During the past decade much has been learned about the diagnosis and management of intersex.[1] New laboratory procedures have made it relatively easy to make a credible diagnosis without exploratory surgery. Sex may now be assigned with confidence fairly soon after birth; corrective surgery and hormone therapy may be carried out later. As a result, intersexes may lead fairly normal lives. Many can be made marriageable; some will be fertile.

MEDICAL ASPECTS OF INTERSEXUALITY

The appropriate sex for an intersex is not always consistent with that established at fertilization or with the type of gonads. Sex in such patients should not be based on any one aspect of sex; all criteria must be considered. In the newborn, special emphasis should be placed on external genital anatomy and on the surgical possibilities of creating functional sex organs. In persons beyond infancy, utmost importance should be attached to the sex that has been adopted; in these cases it is usually best to retain the sex of rearing to prevent psychological maladjustment. Often a person's suitability for a sex may be greatly improved by hormonal therapy and plastic surgery.

As recently as six years ago, most physicians believed that sex of rearing should, almost invariably, be consistent with gonadal sex. This firm belief often resulted in changes of sex well beyond infancy, sometimes in adults. It may be that unreasonable emphasis as to sex is still placed on gonads by some doctors; certainly this idea is prevalent in non-medical groups. It is, of course, desirable to designate the sex indicated by the gonads, but this may not always be reasonable. There need be no hesitation in raising a person contrary to gonadal sex; it is well established that sex orientation develops according to sex of rearing.[2] Physicians must be able to make accurate judgment in such cases, and be prepared to outline the rationale for such decisions. Knowledge of intersexuality is valuable, not only because of possible legal complications, but in order to prevent tragic and embarrassing mistakes.

Before the various forms of intersexuality are described, the criteria of sex that should be used in forming a correct judgment of appropriate sex will be briefly outlined. The criteria are: (1) chromosomal sex, (2) gonadal sex, (3) hormone pattern, (4) internal sex organs, (5) external genitalia, (6) habitus, (7) sex of rearing, and (8) gender role and orientation. In the newborn, the first five criteria are used.

CRITERIA OF SEX

1. *Chromosomal Sex*

An embryo's chromosomal sex is established at fertilization and depends on whether or not an ovum is fertilized by an X or a Y sperm (Fig. 2). Chromosomal females have two X chromosomes, whereas chromosomal males have an X and a Y chromosome. A person's chromosomal sex can be detected by a chromatin test;[3] the most commonly used is the oral smear method.[4, 5] Nuclei of chromosomal females contain a special mass of chromatin, the sex chromatin (Fig 1a),

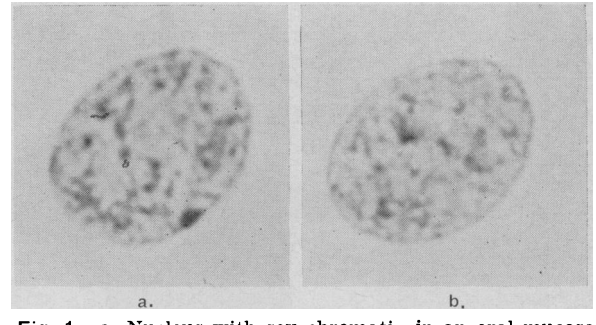

Fig. 1.—a. Nucleus with sex chromatin in an oral mucosal smear from a chromosomal female. b. Nucleus without sex chromatin in an oral smear from a chromosomal male. (Cresylecht violet, × 3000.)

that is not visible in nuclei of chromosomal males (Fig. 1b). The generally accepted hypothesis is that the sex chromatin represents persistent portions of heterochromatic regions of the X chromosomes. Similar parts of the X and Y chromosomes of males do not form a recognizable chromatin mass in interphase nuclei.[6]

If sex chromatin is visible, it may be inferred that the nuclei contain two X chromosomes. Nuclei of this type should be referred to as chromatin-positive, never as female, for such reference could unnecessarily upset a patient if this criterion indicated the sex opposite to the person's rearing. If cells from an intersex are chromatin-positive, it is also likely that two X chromosomes are present, but the actual chromosome constitution cannot be determined by this method. With new cytological techniques it has been shown that intersexes with chromatin-positive nuclei may have the following sex chromosome constitutions: XX, XXX or XXY.[7, 8] It is important to realize also that the

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*This article is based on a paper presented to the Manitoba Medico-Legal Society on November 24, 1959, and published in the *Manitoba Bar News*, 31: 101, 1959. This work is supported by grants from the Committee for Research in Problems of Sex, National Academy of Sciences, National Research Council (U.S.A.), and the National Research Council of Canada.

†Associate Professor of Anatomy, University of Manitoba; Consultant in Anatomy, Children's Hospital, Winnipeg.

‡Recorder, and Assistant Professor, Manitoba Law School.

Canad. M. A. J.
Sept. 24, 1960, vol. 83

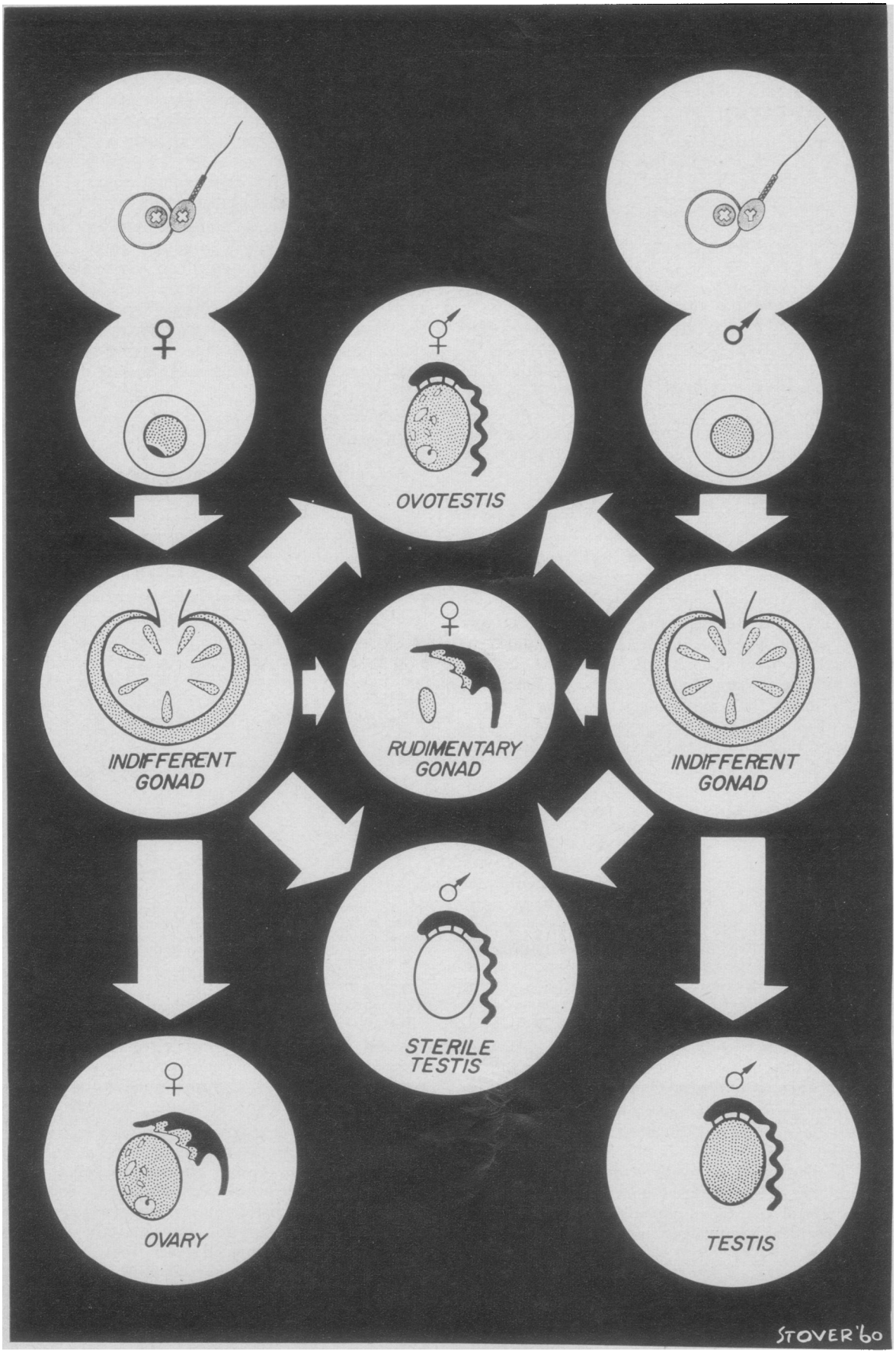

Fig. 2.—Diagrammatic representation of sex determination and differentiation. Normal development is represented in the outside columns; some types of abnormal differentiation are illustrated in the centre column.

chromatin test gives no direct information as to genetic sex; that is, sex gene constitution and arrangement.

If sex chromatin is not visible in a random sample of 100 nuclei, it may be inferred that the XY sex chromosome constitution is present. Cells of this type should be referred to clinically as chromatin-negative. In an intersex this nuclear pattern indicates that two X chromosomes are not present, but it cannot be determined by this method whether the sex chromosome constitution is XY, XO, YO or O. This does not detract from the great practical value of the chromatin test in the differential diagnosis of intersex.[1] Indeed it was the development of a relatively simple method of detecting chromosomal sex that stimulated new interest in intersexuality.[9]

The use of chromosomal sex as the sole criterion of sex would of course be ridiculous. No physician would suggest that a person with the normal appearance of one sex should be raised in the other on the basis of chromosomal sex alone.

2. *Gonadal Sex*

The sex of the gonads apparently depends on the ratio between male-determining and female-determining genes. If the genes for femaleness predominate, ovaries develop, whereas if there is an excess of male-determining genes, testes form. Thus faulty chromosomal distribution and genic abnormalities may cause abnormal gonadal development. Ovaries may develop in chromosomal males, or testes in chromosomal females; these conditions are known to occur in man.[10, 11] In genetic females the cortex of the indifferent gonad differentiates into an ovary and the medulla regresses (Fig. 2); in genetic males, the medulla develops into a testis and the cortex recedes. If there is a genetic disturbance or abnormal hormones are present, both the medulla and cortex may differentiate to form an ovotestis.

Occasionally no gonads form, or more commonly, vestigial structures with ovarian-like stroma develop. This type of gonad may be found in chromosomal males or females, but is commoner in the former.[12] It has been shown that some chromatin-negative patients with this gonadal condition lack a Y chromosome.[13] This and other evidence[14] suggest that the Y chromosome may have an active role in determination of maleness.

3. *Sex Hormone Pattern*

Fetal testes produce hormones or inductor substances that cause masculine differentiation of indifferent sex structures. Failure of testes to form or to produce androgenic hormones results in female development. Thus all fetuses appear feminine, unless masculinizing substances are present.[15] Conversely, masculinization of female fetuses occurs if androgenic substances act on undifferentiated or partially differentiated structures. The common source of such substances in human fetuses is the hyperplastic adrenal. Similar effects may be caused by maternal androgenic substances from masculinizing tumours,[16] or by administered testosterone preparations.[17]

It is particularly important that the hormone status of intersexes be considered. If excessive amounts of androgens are being produced in a female, masculinization will be progressive. At puberty testes in intersexes may produce either androgens or œstrogens; generally if the genitals are ambiguous, testes produce androgens, whereas if the genitals are normal for a female, testes produce œstrogens. Thus gonads in intersex patients may cause embarrassing characteristics to develop at puberty. Signs of contradictory sex development should be watched for around puberty and, if they appear, the condition should be treated before it causes concern.

4. *Internal Sex Organs*

Every fetus has ducts capable of developing into either male or female internal genitalia. If male hormone is present in sufficient quantities at the appropriate time, male organs develop, whereas if it is absent, female organs differentiate regardless of chromosomal or gonadal sex. Consideration of internal genitalia is obviously important for assessment of a patient's marriageability and chances of fertility, but is not of prime importance in assigning sex.

5. *External Genitalia*

By the 9th week of development experts can diagnose sex from external genitalia; the differences are obvious by the 12th week. Masculine genitalia form if adequate male hormone is present during the period of sexual differentiation, regardless of the type of gonads present. If no gonads are present, or no male hormone is produced, female external genitalia develop regardless of genetic sex.

The external genitalia are the criterion commonly used in sex diagnosis; this is certainly the most important criterion in the newborn. The genitals may be unreliable indicators of chromosomal or gonadal sex; often it may be extremely difficult to differentiate a female with a hypertrophied clitoris and rugose labia majora from a cryptorchid hypospadiac with a bifid scrotum.

6. *Habitus*

The development of body form is dependent on genetic and hormonal factors. If intersex patients are diagnosed early and given proper hormonal therapy, the desired appearance can usually be developed. Although changes of sex after infancy are not recommended, contradictory habitus in poorly managed cases is usually the compelling reason for patients to seek a change. Such cases were relatively common a few years ago; there is little excuse for allowing them to occur today.

7. *Sex of Rearing*

After infancy, sex of rearing becomes an exceedingly important criterion of sex. Often by the age of two the social pattern is so well established that a change could cause serious mental confusion. Money *et al.*,[18] after a psychological study of 76 intersexes, concluded that when a change of assignment was made later than early infancy, life adjustment was not significantly improved and was often made worse. Rare voluntary requests for change of sex should be given serious consideration; such a person may have become convinced, because of serious conflict between assigned sex and external genitalia, that a mistake was made at birth. In such instances, after psychiatric consultation, a change may be desirable.

Canad. M. A. J.
Sept. 24, 1960, vol. 83

8. *Gender Role and Orientation*

Good evidence supports the view that psychological masculinity and femininity develop after birth according to experiences of growing up; once firmly established at about 18 months the conviction can rarely be successfully erased.[18] The practical value of this finding is that there need be little fear of psychological maladjustment if it seems appropriate to raise a child opposite to its chromosomal and gonadal sex. Of course development of the desired gender role and orientation in such cases depends upon good clinical management.

DEFINITION OF INTERSEXUALITY

An intersex is a person in whom there is a contradiction of one or more of the following morphological criteria of sex: chromosomal sex, gonadal sex, internal sex organs and external genitalia.

DIAGNOSIS OF INTERSEXUALITY

Most intersexes may be recognized easily at birth because of equivocal external genitalia. Intersexuality is likely to be missed if the genitals appear normal; the condition in such cases is usually detected in adolescents who show abnormal sex development.

The usual diagnostic problem is to differentiate the male pseudohermaphrodite from the female with congenital virilizing adrenal hyperplasia. This may now be done fairly easily by the chromatin test and by determination of urinary 17-ketosteroids. A positive chromatin test and elevated urinary 17-ketosteroids, that can be suppressed by cortisone, establish a diagnosis of congenital adrenogenital syndrome. Female pseudohermaphroditism with non-progressive virilization is strongly indicated if the ketosteroid excretion is normal in a chromatin-positive patient, but the rare condition of true hermaphroditism cannot be excluded unless the gonads have been examined microscopically.

A negative chromatin test in an intersex with ambiguous genitalia usually indicates male pseudohermaphroditism, though true hermaphroditism again cannot be ruled out until the gonads have been examined microscopically.

Intersexes with normal-appearing genitalia are rarely detected in infancy or childhood, unless characteristic physical signs are present or large surveys are conducted.[19] A negative chromatin test in a person with female genitalia suggests gonadal dysgenesis (Turner's syndrome and related conditions)[12] or a type of male pseudohermaphroditism (testicular feminization syndrome).[20] A diagnosis of gonadal dysgenesis is established by the presence of a high titre of urinary gonadotrophin (FSH) and characteristic skeletal defects.[12] Male pseudohermaphrodites with feminizing testes are usually detected when they present with amenorrhœa or inguinal hernia.

A positive chromatin test from a person with male genitalia suggests seminiferous tubule dysgenesis (Klinefelter's syndrome and related conditions)[21] or an excessively masculinized female pseudohermaphrodite. One characteristic feature of seminiferous tubule dysgenesis is an excessive amount of pituitary gonadotrophin (FSH) in the urine; 17-ketosteroid excretion is normal or subnormal. The rarer condition, female pseudohermaphroditism with excessive virilization would be suggested by an increased excretion of 17-ketosteroids in the urine.

POSSIBLE MEDICO-LEGAL PROBLEMS

Assignment of sex in intersexuality being based on several criteria, errors of diagnosis and of judgment may occur. Also, undue emphasis on a criterion of sex (chromosomal, gonadal or hormonal), not essential for social and psychological adjustment, may give rise to differences of opinion as to sex. Those directly concerned with problems of sex have often wondered whether medical and legal opinion would differ greatly concerning modern biological views of sex. It was this concern, sparked by curiosity, that prompted consideration of the medico-legal aspects of intersexuality. Illustrative cases will be described to raise possible questions.

Males with Criteria of Female Sex

CASE 1.—Jones and Scott[22] described a fairly normal-appearing boy who was diagnosed as a female pseudohermaphrodite with congenital excessive virilizing adrenal hyperplasia. The patient was designated a male at birth because the genitalia were predominantly masculine. Investigation five years later revealed ovaries, tubes and a uterus. The criteria of sex were as follows:

TABLE I.

Criteria of sex	*Male*	*Female*
Chromosomal sex		*
Gonadal sex		*
Sex hormone pattern	*	
Internal sex organs		*
External genitalia	*	
Habitus	*	
Assigned sex	*	
Gender role and orientation	*	

Criteria of maleness predominate because he was assigned as a boy and developed a male sex role.

This case illustrates the prime importance of external genital anatomy in sex assignment at birth. Although female internal genitalia were not suspected at birth, this knowledge probably would not have affected the designation. To adapt this boy further to the male sex, the internal genitalia and ovaries were removed. Medically there is no doubt about the appropriate sex for this patient.

CASE 2.—Money *et al.*[25] described a young married man who was a female pseudohermaphrodite with non-progressive virilization. The external genitalia were equivocal; a uterus, tubes and ovaries were present; breasts and feminine habitus had begun to develop at puberty. After removal of contradictory sex structures, plastic repair of the external genitalia and hormone therapy, the young man had married and was well adjusted. The criteria of sex, before treatment, were as follows:

TABLE II.

Criteria of sex	*Male*	*Ambiguous*	*Female*
Chromosomal sex			*
Gonadal sex			*
Sex hormone pattern			*
Internal sex organs			*
External genitalia		*	
Habitus			*
Assigned sex	*		
Gender role and orientation	*		

It is obvious that a misdiagnosis was made at birth, but at the time there was no practical way of detect-

ing chromosomal sex. In view of the ambiguous external genitalia, the error was understandable. There would be no excuse for misdiagnosis in such a case today. This case illustrates how well such patients can become adjusted in the "wrong" sex if they are properly managed. It would have been ridiculous to alter this patient's sex at puberty, as he had developed an unchangeable masculine gender role and orientation. Thus, medically this person is a male, despite outright contradiction by five criteria of sex.

CASE 3.—A patient with seminiferous-tubule dysgenesis associated with Klinefelter's syndrome was studied here recently. He had gynæcomastia, eunuchoidism, a slightly female body contour, a small penis, no beard, and a high-pitched voice. The criteria of sex were as follows:

TABLE III.

Criteria of sex	*Male*	*Ambiguous*	*Female*
Chromosomal sex			*
Gonadal sex	*		
Sex hormone pattern		*	
Internal sex organs	*		
External genitalia	*		
Habitus		*	
Assigned sex	*		
Gender role and orientation	*		

Clinically and socially this patient is a male. Although he has several feminine characteristics, he has only one morphological criterion of femaleness. Many such patients have XXY sex chromosomes, thus they are not really chromosomal females.

CASE 4.—Armstrong[26] described a male patient with transvestism, a strong desire to wear female clothing and to be accepted as a female. The patient's external genitalia were masculine; his habitus was fairly feminine, mainly because he took female hormones; his mannerisms, facial expressions and voice were feminine. All morphological criteria of sex except habitus indicated maleness, and the female habitus was produced by self-administered hormones. The patient's gender role and orientation were female.

This patient is not an intersex by our definition, as there is only contradiction between physical and psychological criteria of sex. It is this type of patient that is publicized so much; for example, "Father of Two Becomes Woman". It is well known that in Denmark selected patients are emasculinized by castration and by amputation of the penis, and then feminized by plastic construction of female-like external genitalia and hormone therapy. What is the person's sex now?

The criteria of sex in such a person would be as follows:

TABLE IV.

Criteria of sex	*Male*	*Female*
Chromosomal sex	*	
Gonadal sex	castrated	
Sex hormone pattern		hormonally created
Internal sex organs	*	
External genitalia		surgically created
Habitus		hormonally created
Assigned sex	*	
Gender role and orientation		*

Only one of the four criteria indicating femaleness developed naturally.

If such a person's sex were changed, criteria of the female would predominate. Would this make him a female? The answer is "no" clinically and probably socially also. He is a mutilated, non-marriageable eunuch who will probably not be accepted by either sex. If accepted as a female legally, embarrassing social and legal problems might ensue.

Females with Criteria of Male Sex

CASE 1.—A happily married woman studied because of tiredness and amenorrhœa had good breast development, a normal vagina and normal external genitalia.[23] Testes and vestigial female internal organs were discovered at laparotomy. The diagnosis was testicular feminizing syndrome (male pseudohermaphoditism); the criteria of sex were as follows:

TABLE V.

Criteria of sex	*Male*	*Female*
Chromosomal sex	*	
Gonadal sex	*	
Sex hormone pattern		*
Internal sex organs		*
External genitalia		*
Habitus		*
Assigned sex		*
Gender role and orientation		*

This woman is unequivocally female by clinical and social standards.

Patients with gonadal dysgenesis and chromatin-negative nuclei would show similar criteria of sex, and similarly there would be no doubt about their medical sex.

CASE 2.—Brewer, Jones and Culver[27] reported a case of true hermaphroditism that had an unusual feature. The certification of birth stated that the patient was male. However, the mother did not accept the doctor's decision and raised her baby as a girl. The infant developed into an attractive woman: breast development was good, the habitus definitely feminine. She became concerned about her genitalia when marriage was proposed: the genitals were ambiguous but more masculine than feminine. Surgical exploration revealed ovotestes, a uterus, tubes and a vagina. The criteria of sex were as follows:

TABLE VI.

Criteria of sex	*Male*	*Ambiguous*	*Female*
Chromosomal sex		*not determined*	
Gonadal sex		*	
Sex hormone pattern			*
Internal sex organs			*
External genitalia	*		
Habitus			*
Assigned sex	*		
Gender role and orientation			*

Obviously the patient is female clinically. What is her legal sex, the sex on her birth certificate or the sex the mother decided?

CASE 3.—A 15-year-old girl with male pseudohermaphroditism was described by Jones and Scott.[24] She had testes, female genitalia with a large phallus and a vagina 5 cm. long; no other female organs were found. The criteria of sex were as follows:

Canad. M. A. J.
Sept. 24, 1960, vol. 83

TABLE VII.

Criteria of sex	*Male*	*Female*
Chromosomal sex	*	
Gonadal sex	*	
Sex hormone pattern	*	
Internal sex organs	predominantly	
External genitalia		predominantly
Habitus	predominantly	
Assigned sex		*
Gender role and orientation		*

This patient is a female clinically because of her feminine gender role and orientation, and predominantly female external genitalia.

Nowadays similar patients, known to be chromosomal males at birth, are also raised as females, as their genitalia are more suited for that sex. Reconstruction of the genitalia and removal of contradictory structures enable such persons to lead fairly normal lives as females. In the past such females were usually not suspected of having testes until they became extremely masculinized. Many tragic cases could be cited. One will illustrate the consequences.

CASE 4.—A young married woman became concerned about her masculine habitus, her sexual inadequacy, and her strong affection for members of her sex. On examination it was found that she was actually a male with perineal hypospadias. In view of the patient's masculine orientation and masculine organs, the physician recommended that she change her sex. The change of registration and annulment of marriage were carried out without legal complications.

This case illustrates that in rare instances sex should possibly be changed if satisfactory psychological adjustment may be expected. This patient could have remained a female and her role improved by castration, plastic surgery and hormone therapy. But, for reasons previously stated, it was considered best to change "her" sex. After repair of the hypospadias the patient appeared to be well adjusted.

It should be clear now that sex should not be designated by any one criterion; several should be considered. The relative value of each depends on the particular case, with regard to age, degree of psychological adjustment, surgical possibilities for reconstruction, etc. The modern trend is to make an early decision based on all available evidence: often this is a matter of clinical experience. Assignment of sex in such cases should be considered carefully, but an unequivocal decision should be made as soon as possible after birth.

Many possible questions with legal implications may be raised concerning persons in whom there is a contradiction of criteria of sex: Could such persons consummate a marriage? Would they create problems in the field of wills and inheritance? Could such persons, claiming that they were not real males or females, escape charges of rape, indecency and adultery? The answers to these and other questions will become obvious after the next section (Part II) has been read.

GENERAL PRACTICE

THE GENERAL PRACTITIONER AS A SPECIALIST

W. VICTOR JOHNSTON, M.B.*

SOME CANADIAN medical educationalists have computed that it costs about $30,000 to produce a general physician and $50,000 to produce a specialist. These figures seem to reflect the relative importance which is placed by educationalists on the preparation for medicine as practised by general physicians and specialists. Many factors have contributed to the development of this situation in medical education. We submit that the time is rapidly approaching when as much thought, planning, and resulting expense should be devoted to the training of general practitioners as to that of specialists.

With increasing rapidity there is emerging a new concept of the general practitioner trained to take a total approach to all the common problems of his patients and, what is just as important, trained to work in reciprocal cooperation with highly competent specialists.

It is becoming increasingly clear that the majority of people want a personalized type of medical care, in spite of all factors tending to depersonalize it. They want it to be warmly human as well as scientific. They want their physician to bring the best technical knowledge as well as the priceless remedy of a personal interest in them as people. They wish an emergency service at all times, as serious illness knows no convenient hours, and this requires that the doctor must be interested in making house calls.

The idea that a group of specialists can replace good general physicians has been advocated for years but there is increasing evidence that this is too expensive, cumbersome and inefficient. The myth that a general practitioner cannot perform the common tasks of medicine as well as specialists can be easily dispelled by a thorough study of good general practice conducted by well-trained general physicians and backed by a consulting group of specialists.

There is a very articulate school of medical opinion that overemphasizes to the general practitioner the increasing complexity of modern medicine. In this, they are doing a disservice to medicine, where

*Executive Director, College of General Practice (Medicine) of Canada.